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Acute esophageal necrosis masquerading acute coronary syndrome

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Abstract

Acute esophageal necrosis (AEN) also known as "black esophagus" or "acute necrotizing esophagus" is a rare entity characterized by striking endoscopic findings of circumferential black coloring of the esophagus. AEN most frequently seen in the distal esophagus and can extend proximally along the entire esophagus. Characteristically, the circumferential black mucosa stops abruptly at the EGJ. AEN tends to present as acute upper gastrointestinal bleeding, though other symptoms including dysphagia and epigastric pain have been described. The etiology of AEN is multifactorial including a combination of ischemic insult, mucosal barrier defect, and a backflow injury of gastric secretions. Described is a case of AEN in a patient with history of uncontrolled diabetes who presented with an atypical chest pain mimicking acute coronary syndrome with negative subsequent cardiovascular workup.

Keywords
Esophageal necrosis, Acute coronary syndrome, Chest pain

INTRODUCTION

Acute esophageal necrosis (AEN) also known as "black esophagus," "acute necrotizing esophagus," or "Gurvits syndrome" is a rare syndrome with characteristic endoscopic finding of circumferential black appearance of the esophagus beginning at the gastroesophageal junction (GEJ) and extending proximally at variable lengths.[1-4] Prevalence of AEN is estimated to be anywhere from 0.001 to 0.2% in endoscopy series.[5,6] AEN is four times more prevalent among males than females and with a peak incidence in the sixth decade of life.[6,7]

Most patients with AEN present with an acute upper gastrointestinal bleeding, though other symptoms such as dysphagia, epigastric pain, and chest pain have been described.[6,8] Complications range from stenosis or stricture formation at the distal esophagus to perforation and mediastinitis. Herein, we present a case of isolated AEN in a patient with solely uncontrolled diabetes and a negative cardiovascular workup.

CASE REPORT

An 80-year-old male with past medical history of hypertension, dyslipidemia, and diabetes mellitus non-compliant to the prescribed medications, presented to the emergency department with 1-hour complaint of right-sided chest pain that was described as aching, non-radiating, without any aggravating or relieving factors, 10/10 in intensity, and was accompanied by nausea and two episodes of maroon-colored emesis. The patient denied any ingestion of toxic substances and any prior similar episodes. On presentation, the patient was afebrile, and blood pressure of 109/45,

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heart rate 75, respiratory rate of 18, and room air oximetry of 98%. Physical examination, including fundoscopy was unrevealing. Labs workup revealed serum sodium of 125 mEq/L [135-145 mEq/L], potassium 5.1 mEq/L [3.5-5.0 mEq/L], chloride 91 mEq/L [98-107 mEq/L], bicarbonate 24 mEq/L [21-24 mEq/L], urea 145,7 mg/dL [14.9 – 51.4 mg/dL], creatinine 2.95 mg/dL [0.6-1.30 mg/dL], blood glucose 492 mg/dL [70-110], WBC 13.5 [4.5-11.0 × 10^3/mm^3], Hb 12.3 g/dL [13.5-17.5g/dL], platelets 242,000/mm^3 [140k-440k/mm^3], BNP 154 pg/mL [125-450 pg/mL], and troponin 0.062 ng/mL [0.01-0.30 ng/mL]. Electrocardiogram showed normal sinus rhythm at the rate of 94 beats per minute without ST or T-wave abnormalities. Morphine was administered along with nitroglycerine for the chest pain without any improvement in symptoms. Therefore, given the persistent chest pain and concern for non-ST-segment elevation myocardial infarction, the patient was started on a heparin drip and antiplatelet therapy in the emergency department. Cardiology was consulted who performed a cardiac catheterization which did not show any critical stenosis with an estimated ejection fraction of 55%. They recommended medical therapy including anti-hypertensives, statin, and blood glucose control. The computed tomography of the abdomen and pelvis without contrast was unremarkable. Gastroenterology was then consulted due to abdominal symptoms of nausea, vomiting, and hematemesis. Patient underwent an esophagogastroduodenoscopy (EGD), which showed circumferential, black-appearing, necrotic esophageal mucosa without active bleeding extending from the mid to distal esophagus (Figure 1). There was also mild chronic gastritis without any

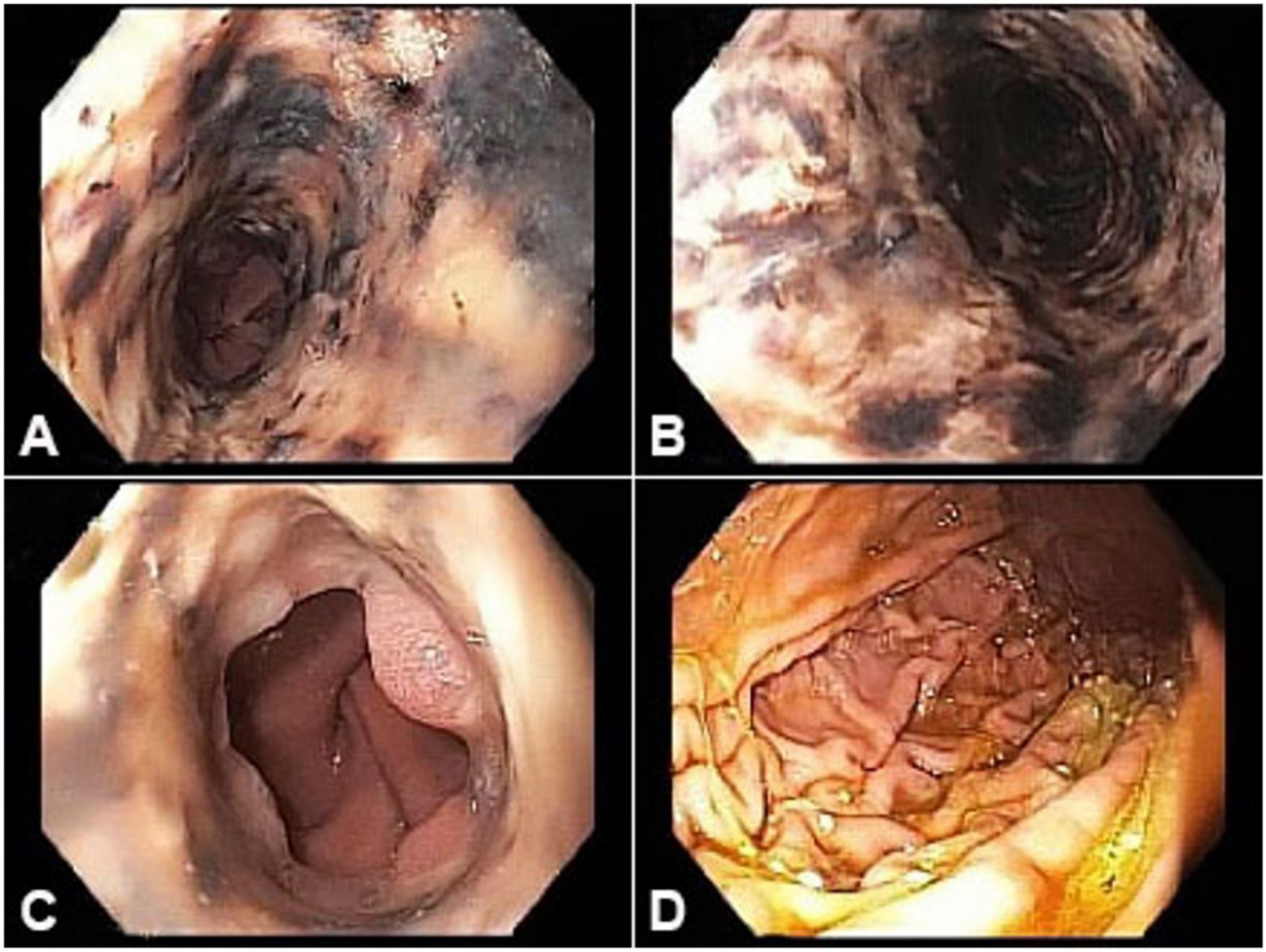

Figure 1. Endoscopic images showing: **A** and **B -** the mid to distal esophagus with circumferential, black-appearing, necrotic esophageal mucosa without active bleeding; **C -** the gastroesophageal junction with normal healthy appearing gastric mucosa; **D -** the gastric body mucosa.

evidence of active bleeding. The patient was treated with a proton pump inhibitor and sucralfate. Patient had an uneventful hospital course lasting 5 days and was discharged with close follow up.

DISCUSSION

Acute esophageal necrosis (AEN) is an under-reported condition described by endoscopic findings of circumferential black appearance of the esophagus extending distally to the gastroesophageal junction (GEJ). The reason for such characteristic finding is attributed to the reduced vascularity of the distal esophagus.[1,2] The etiology is thought to be multifactorial including a combination of ischemic insult, mucosal barrier defect, and a backflow injury of gastric secretions.[1,4,9-11] There are certain conditions which predispose patients to AEN such as male sex, older age, and chronic comorbidities like diabetes mellitus, hematologic malignancies, malnutrition, renal insufficiency, cardiovascular insufficiency, trauma, and thromboembolic phenomena.[1,4,7-11] AEN typically presents with an acute upper gastrointestinal bleeding, though other symptoms such as dysphagia, epigastric pain, and chest pain have been described.

In the case of our patient, severe and relentless chest pain along with episodes of emesis were the chief complaints. Given his history of uncontrolled diabetes, hypertension, and dyslipidemia, the severe chest pain symptoms were initially attributed to a likely cardiovascular event i.e. non-ST-segment myocardial infarction (NSTEMI), despite the absence of prior symptoms of chest pain or angina pectoris. However, severe chest pain with vomiting, in the absence of significant EKG findings, or significant elevation of troponins should have provided a clue to consider other differentials aside from acute coronary syndrome. Additionally, administration of adequate narcotic analgesics failed to subside the pain symptoms, further pointing to a source other than a cardiovascular event for the symptoms.

Gastrointestinal conditions frequently masquerade as cardiac symptoms; therefore, it is important to keep in mind esophageal etiologies when a cardiac workup is negative. Thinking of an esophageal cause for the source of chest pain would have prevented the patient from being subjected to unnecessary and time-consuming initial cardiac work-up. Furthermore, a prompt upper endoscopy is vital to classify the degree of esophageal necrosis, as the prognosis and treatment options may vary. The reported mortality depending on the underlying comorbidities is about 32%; with 6% of deaths directly related to the complications of acute esophageal necrosis.[1-4] Initial treatment lies in supportive measures including adequate intravenous hydration, correction of anemia, esophageal rest by nil-per-os, and management of underlying medical conditions. After endoscopy, medical management includes proton-pump inhibitors, sucralfate, and empiric antibiotics in cases of complications. Surgical intervention is usually reserved for individuals with esophageal perforation and subsequent mediastinitis and abscess.

AEN is a rare, grave condition that should be considered as a differential in those individuals with a predisposition and negative cardiac workup.

REFERENCES

1. Gurvits GE. Black esophagus: acute esophageal necrosis syndrome. World J Gastroenterol. 2010;16(26):3219-25. http://dx.doi.org/10.3748/wjg.v16.i26.3219. PMid:20614476.
2. Shafa S, Sharma N, Keshishian J, Dellon ES. The black esophagus: a rare but deadly disease. ACG Case Rep J. 2016;3(2):88-91. http://dx.doi.org/10.14309/crj.2016.9. PMid:26958555.
3. Sako A, Kitayama J, Inoue T, Kaizaki S, Nagawa H, Suzuki H. Black oesophagus: cause. Gut. 2005;54(2):192-227. http://dx.doi.org/10.1136/gut.2004.044784. PMid:15647179.
4. Gurvits GE, Shapsis A, Lau N, Gualtieri N, Robilotti JG. Acute esophageal necrosis: a rare syndrome. J Gastroenterol. 2007;42(1):29-38. http://dx.doi.org/10.1007/s00535-006-1974-z. PMid:17322991.
5. Postlethwait RW, Musser AW. Changes in the esophagus in 1,000 autopsy specimens. J Thorac Cardiovasc Surg. 1974;68(6):953-6. PMid:4420581.
6. Lacy BE, Toor A, Bensen SP, Rothstein RI, Maheshwari Y. Acute esophageal necrosis: report of two cases and a review of the literature. Gastrointest Endosc. 1999;49(4 Pt 1):527-32. http://dx.doi.org/10.1016/S0016-5107(99)70058-1. PMid:10202074.
7. Gurvits GE, Cherian K, Shami MN, et al. Black esophagus: new insights and multicenter international experience in 2014. Dig Dis Sci. 2015;60(2):444-53. http://dx.doi.org/10.1007/s10620-014-3382-1. PMid:25297468.

8. Soussan EB, Savoye G, Hochain P, et al. Acute esophageal necrosis: a 1-year prospective study. Gastrointest Endosc. 2002;56(2):213-7. http://dx.doi.org/10.1016/S0016-5107(02)70180-6. PMid:12145599.

9. Goldenberg SP, Wain SL, Marignani P. Acute necrotizing esophagitis. Gastroenterology. 1990;98(2):493-6. http://dx.doi.org/10.1016/0016-5085(90)90844-Q. PMid:2295407.

10. Jacobsen NO, Christiansen J, Kruse A. Incidence of oesophageal necrosis in an autopsy material. APMIS. 2003;111(5):591-4. http://dx.doi.org/10.1034/j.1600-0463.2003.1110509.x. PMid:12887512.

11. Forster R, Durso DA, Vattimo EFQ, et al. Black esophagus: exploring the dark. Autops Case Rep. 2013;3(3):41-8. http://dx.doi.org/10.4322/acr.2013.027. PMid:31528617.

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